Pregnancy with Essential Thrombocythemia: A Case Report

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SUMMARY

Objective: We describe a woman with essential thrombocythemia (ET) who experienced a successful pregnancy with no specific treatment.

Case Report: A 30-year-old primiparous female was incidentally found to have ET with no specific symptoms at a gestational age (GA) of 8 weeks. Initially, her platelet count was 1,100 × 10^9/L, which declined to a median of 678 × 10^9/L (range, 663–753 × 10^9/L) during the second and third trimesters, without treatment. A healthy male baby weighing 3,330 g was born by cesarean delivery due to breech presentation at a GA of 38 weeks. The pre-delivery platelet count was 605 × 10^9/L. No bleeding or major thromboembolic complications were encountered in the peripartum period. The maternal platelet count returned to 1,230 × 10^9/L and 1,320 × 10^9/L, respectively, at 2 months and 3 years after delivery. The child had normal physical and mental development at 2 years and 9 months old.

Conclusion: Management of ET during pregnancy is still controversial. Platelet count, either at diagnosis or at conception, fails to predict the pregnancy outcome. The treatment plan should be based on clinical judgment that weighs the risk of thrombohemorrhagic complications against related morbidity and side effects. [*Taiwanese J Obstet Gynecol* 2004;43(4):237–239]

Key Words: essential thrombocythemia, pregnancy

Introduction

Essential thrombocythemia (ET) is a rare myeloproliferative disease manifesting an elevated platelet count and megakaryocytic hyperplasia. The diagnosis is made after the exclusion of secondary thrombocytosis (Philadelphia chromosome negative, normal red blood cell mass, normal iron status, absence of chronic infection and no systemic disease or malignancy), following the criteria of the Polycythemia Vera Study Group [1]. The median age at diagnosis is approximately 60 years, and as many as 20% of patients are diagnosed before the age of 40 years. It is more common in women than in men (female:male, 1.6:1). The clinical manifestations of ET include vasomotor disturbances (headaches, dizziness, acral dysesthesia, visual symptoms or livedo reticularis), thrombohemorrhage, frequent miscarriages during the first trimester and, rarely, transformations into acute myeloid leukemia or myelofibrosis with myeloid metaplasia, although one-third of patients are asymptomatic. There is no current consensus on the optimal management of ET during pregnancy. Management varies from no treatment, through anti-platelet aggregants (aspirin and/or heparin) to the use of platelet apheresis or cytoreductive therapy (interferon-alpha [IFN-α], hydroxyurea, busulfan or anagrelide). Obstetric outcomes are variable, ranging from early miscarriage to normal term delivery.

We report the case of a pregnant woman with ET and the associated clinical course and pregnancy outcome.

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Case Report

A 30-year-old primiparous woman at a gestational age (GA) of 8 weeks was referred from a local clinic because of thrombocytosis (platelet count, 1,100 × 10^9/L) on routine complete blood analysis. She was asymptomatic, and a survey of possible etiologies of thrombocytosis was performed. The Cr-51 red cell volume study showed normal red cell mass. The leukocyte alkaline phosphatase score was within the normal range. Bone marrow biopsy over the posterior iliac crest favored ET. Upper abdominal sonography revealed neither hepatomegaly nor splenomegaly. Thus, a diagnosis of ET was established according to the criteria defined by the Polycythemia Vera Study Group [1]. No specific treatment was given on the recommendation of the hematologist, and the platelet count was routinely checked every 4 weeks. The platelet count declined to a median of 678 × 10^9/L (range, 663–753 × 10^9/L) during the second and third trimesters. Serial ultrasound examination revealed normal fetal growth and normal umbilical arterial resistance. The pregnancy course was uneventful. A healthy male baby weighing 3,330 g was born by cesarean delivery due to breech presentation at a GA of 38 weeks. The maternal platelet count immediately before delivery was 605 × 10^9/L. No bleeding or major thromboembolic complications were encountered in the peripartum period. The platelet count was 1,230 × 10^9/L and 1,320 × 10^9/L at 2 months and 3 years, respectively, after delivery, despite no treatment (Figure). The child had normal physical and mental development at 2 years and 9 months old.

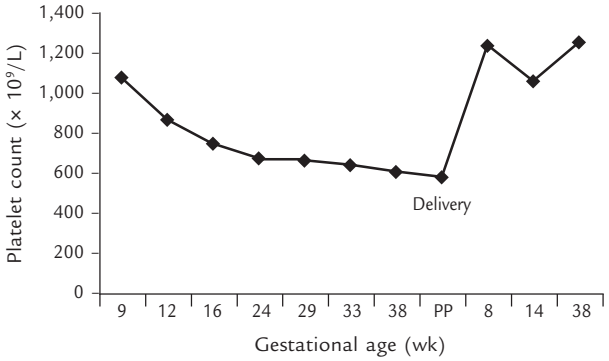

Figure. Platelet count during pregnancy and postpartum (PP).

Discussion

Among pregnancies with ET, 50.3% result in normal babies and 25.9% in spontaneous abortions (mostly in the first trimester) [2]. A trend towards a decrease in the platelet count is observed in most cases of ET during pregnancy, even without treatment. Mechanisms for this include increased blood volume, higher concentration of estrogen and adrenocortical hormones, release of IFN-like factor from the placenta or fetus that downregulates platelet production, and a cytokine-mediated decrease in megakaryocyte number.

Therapy with aspirin may reduce the risk of spontaneous abortion in the first trimester and intrauterine fetal death when compared with no treatment, although thrombotic issues and bleeding during the postpartum period do not significantly differ between patients with and without aspirin treatment [2]. Although cytoreductive therapy during pregnancy may increase the possibility of teratogenicity, IFN-α seems to be safer than hydroxyurea. Amniotic fluid always contains significant amounts of IFN-α, and a physiologic role in the growth and development of the normal placenta and in the regulation of the maternofetal graft interaction has been postulated [3]. This suggests that exogenous IFN might not be harmful to the fetus.

Platelet apheresis can be used in acute situations such as massive thrombosis or hemorrhage with no risk to the fetus, but there are disadvantages, including the expense, the amount of time consumed and the rapid return to pretreatment levels. In addition, no significant correlation has been shown between the thrombotic risk and platelet count.

The platelet count, either at diagnosis or at conception, clearly does not predict pregnancy outcome with ET [4]. Thus, low-risk asymptomatic patients who have no history of thrombosis are usually managed by observation alone, regardless of the degree of thrombocytosis. In contrast, high-risk patients aged more than 60 years or who have a history of thrombosis may benefit from cytoreductive therapy [5]. We conclude that the optimal management of ET in pregnancy is still debatable. The treatment plan should be based on clinical judgment that weighs the risk of thrombohemorrhagic complications against the related morbidity and side effects.

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